matic since age 15, and did not complain of bone pain.

Physical examination revealed a normal intrauterine pregnancy at about five and one-half months gestation. The sclerae were blue and the forehead showed frontal bone prominence. The patient's development otherwise appeared normal, and she walked easily without limp or deformity. Range of motion of all joints was normal. Pelvic examination showed clinically adequate pelvic dimensions. Routine laboratory studies were within normal limits except for hematocrit of 34 per cent.

The patient was treated with iron by mouth and was observed through an uneventful prenatal course. Weight gain was 16 pounds. Results of x-ray pelvimetry in February 1966 were within normal limits and there were no apparent fetal abnormalities. The patient's bones were noted to be osteoporotic. In March 1966 a scout film revealed no fetal fractures. After spontaneous nine-hour labor at term, a normal vaginal delivery of a seven pound ten and three-fourths ounce girl was carried out. A right mediolateral episiotomy and pudendal block anesthesia were used. The patient tolerated the dorsal lithotomy position during delivery without difficulty. There was no excess bleeding or any other postpartum complications. X-ray examination of the pelvis after delivery showed no fractures.

The infant had an Apgar score of 9 and showed no abnormalities on physical examination except for slightly bluish sclerae. There was no evidence of any fractures or deformities, and muscle tone was normal. She was followed until seven months of age, and development was normal to that time in all respects.

Discussion

Management of pregnancy and labor in patients with osteogenesis imperfecta is very similar to that of a normal situation.[1] There is no special indication for interruption of pregnancy in a patient with this disease. There seems to be a somewhat greater need for administration of iron by mouth in the prenatal period. X-ray films should be taken late in pregnancy to evaluate pelvic dimensions and possible fetal fractures. To reduce the risk of fractures, special care should be taken in positioning the patient for delivery, and x-ray films of the maternal pelvis should be made in the postpartum period. Indications for vaginal delivery or cesarian section are the same as in any other pregnancy. Osteogenesis imperfecta congenita can be easily recognized at birth by deformity of extremities or softness and pliability of the cranium, and often the infant is stillborn.[2,5]

The genetic basis for this disease is not firmly established. In one series of 16 infants with osteogenesis imperfecta congenita, 12 had no family history of the disease.[2] A postulation of dominant genetic transmission is favored, but genetic "penetrance" is often so slight that descendants are not affected; unaffected children of an affected parent are not thought to transmit the disease.[8] Sporadic cases are probably mutative.[6]

Summary

A case of pregnancy in a primigravida with osteogenesis imperfecta is presented, together with a discussion of the disease and its management from an obstetrical point of view.

REFERENCES

1. Cohn, S. L., Schreier, R., and Feld, D.: Osteogenesis imperfecta and pregnancy—Report of a case, Obst. & Gynec., 20:107-108, July 1962.
2. Freda, V. J., Vosburgh, G. J., and DiLiberti, C.: Osteogenesis imperfecta congenita—A presentation of 16 cases and review of the literature, Obst. & Gynec., 18: 535-547, November 1961.
3. Heys, F. M., Blattner, R. J., and Robinson, H. B.: Osteogenesis imperfecta and odontogenesis imperfecta: Clinical and genetic aspects in eighteen families, J. Ped., 56:234-245, February 1960.
4. Levin, E. J.: Osteogenesis imperfecta in the adult, Am. J. Roentgen., 91:973-978, May 1964.
5. Mussio, T. J.: Osteogenesis imperfecta congenita—Report of a case discovered in utero, Obst. & Gynec., 15:361-363, March 1960.
6. Reed, S. C.: Counseling in Medical Genetics, W. B. Saunders Company, Philadelphia and London, p. 243, 1955.
7. Sarma, V.: A case of intrauterine osteogenesis imperfecta, British Med. J., 5216:1856-1857, December 1960.
8. Stool, N., and Sullivan, C. R.: Osteogenesis imperfecta in 95 year old woman, Proc. Mayo Clinic, 34:523-528, 1959.
9. White, C. A.: Osteogenesis imperfecta tarda and pregnancy, Obst. & Gynec., 22:792-794, December 1963.

Atypical Bilateral Temporal Artery Disease

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Diagnosis of temporal giant cell arteritis in patients less than 48 years of age is uncommon,[4] but examination of sections in random post mortem

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examinations showed it to be more common than suspected.[1] It may be an occult lesion,[7] or the typical histologic phenomena may occur in a vessel other than the temporal artery.[6] In the case here reported, a 38 year old man had a classical history of the disease but the histopathological findings were unusual. Biopsy of a specimen taken from the left temporal artery showed arteritis without necrosis or giant cells. Eight months later a specimen from the opposite temporal artery showed a dissecting aneurysm.

Report of a Case

A 38-year-old caucasian man was admitted for evaluation to Highland General Hospital. He had first been seen in the Emergency Room in April 1965 with first and second degree burns over the face and hands from a gasoline fire. He was treated with nitrofurazone (Furacin®) dressings and in a month recovered.

At a follow-up visit to the outpatient clinic in May 1965, he first complained of headaches which had begun several weeks before the burn. This pain was located in a 6 cm diameter area in the left temporal region, with radiation to the left temporomandibular joint and anteriorly over the left side of the forehead. The pain was severe, throbbing and lancinating in character, occurred two to three times daily, lasted 15 to 30 minutes, and was relieved by taking aspirin. There was no history of aura, nausea, vomiting, lacrimation, rhinorrhea or allergic disease. In January 1965, however, the patient had noticed episodic facial flushing which, on questioning, he recalled might have been simultaneous with headaches. Administration of aspirin with codeine 30 mg as needed for pain and of phenobarbital 15.0 mg four times a day was begun.

The medical history elicited congenital external strabismus with associated paretic nystagmus. There was no history of familial disease. The patient smoked one to two packages of cigarettes daily and consumed ethanol in small amounts.

In August 1965 the severity of the pain increased and the temporal area became sore. The left temporal artery was swollen and tender. A biopsy specimen was taken. It showed arteritis without necrosis or giant cells (Figure 1.)

After biopsy, prednisone was given, 30 mg daily. Headaches temporarily diminished after the excisional biopsy, but then recurred, and meprobamate, 1.2 gm daily, was added to the regimen. There was a slow remission over the subsequent months.

In February 1966 mild acne vulgaris developed which was treated with improved skin hygiene and baths with a hexachlorophene emulsion. The patient also said that facial flushing had become virtually constant but still fluctuated somewhat with severity of head pain.

In March he first complained of right temporal headaches associated with tenderness over the temporal artery. These headaches were similar to those on the left side and a specimen was excised from the right temporal artery for biopsy. Headaches persisted without remission after operation and the previously prescribed drugs were continued—prednisone 30 mg daily, meprobamate 1.2 gm daily, aspirin with codeine 30 mg as needed, and antacids.

The patient was again admitted to hospital and on physical examination pronounced erythema of the face and neck was noted and there was decided venous telangiectasis of the forehead and nose, acne vulgaris of the face, back and chest, and external strabismus with horizontal nystagmus on the right. The head was normal in shape and size. In the temporal regions there were two well-healed biopsy sites. Over the remaining right temporal artery there was a slightly tender area 2 cm in diameter. The retinal arterioles were slightly narrowed. Peripheral pulses were normal. There were no subcutaneous nodules or splinter hemorrhages and no enlargement of lymph nodes. Blood pressure on repeated readings averaged 110/80 mm of mercury. On a few occasions the diastolic pressure was 100 mm.

Packed cell volume was 44 per cent and leu-

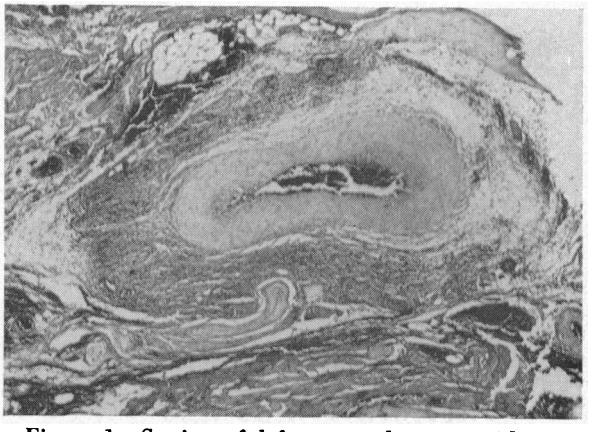

Figure 1.—Section of left temporal artery with periadvential inflammatory exudate. Media is intact. (Hematoxylin and eosin stain, ×40.)

kocytes numbered 13,600 cu mm with segmented neutrophiles 64 per cent, band forms 4 per cent, lymphocytes 31 per cent, and eosinophiles 1 per cent. The erythrocyte sedimentation rate (Westgren) was 9 mm in one hour. Two L.E. cell preparations were negative. The platelet count was 256,000 per cu mm. A Venereal Disease Research Laboratory (VDRL) test was non-reactive. Results of urinalysis were within normal limits. Blood urea nitrogen was 13 mg per 100 ml. Serum albumin was 4.32 gm and globulin was 1.83 gm per 100 ml. A liver profile was entirely within normal limits. Catecholamines in a 24-hour specimen of urine were 30 mcg (normal under 180 mcg) and 5-hydroxyindolacetic acid was 9.7 mg (normal 0.5 to 7.0 mg). X-ray films of the chest and skull and an electrocardiogram were all within normal limits. Biopsy of the skin and the gastrocnemius muscle biopsy showed no pathologic changes.

In the hospital the patient had episodes of severe right temporal headaches with radiation over the right side of the forehead. These occurred three to six times daily, lasting 15 to 60 minutes and were poorly relieved with analgesics. An attempt to decrease the dose of prednisone was initiated and the patient was discharged to clinic care.

Patholigist's Report

The specimen from the left temporal artery was 0.4 cm in diameter with a small lumen. The wall appeared uniformly thickened but elastic. The right temporal artery was 0.5 cm in diameter. On section the wall was dark red. The lumen was not seen.

On microscopic examination there appeared to be no thrombotic occlusion of the lumen of the left temporal artery (Figure 2). A slight focal subintimal thickening of the wall was noted. The elastica and media were intact. In adventitia and surrounding connective tissue there was dense lymphocytic inflammatory infiltration with many scattered eosinophils. No giant cells were seen. A few scattered eosinophils and lymphocytes were present in the subintimal thickening.

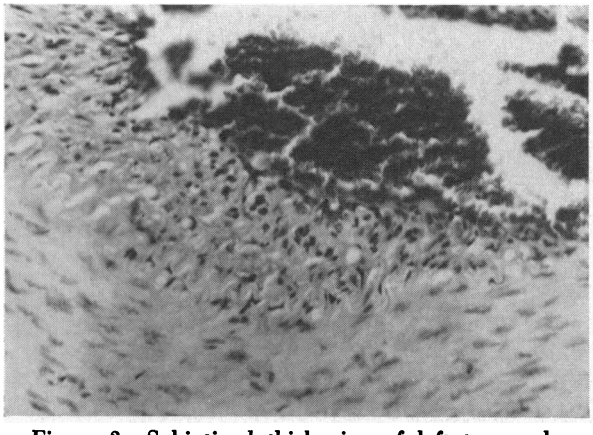

Figure 2.—Subintimal thickening of left temporal artery with few inflammatory cells. (Hematoxylin and eosin stain, × 250.)

In the right temporal artery (Figure 3) a dissecting channel of blood penetrated through the intima and media. The aneurysmal sac was limited by a thin layer of adventitia. The periadventitial connective tissue contained lymphocytes and few polymorphonuclear leukocytes. This inflammatory reaction was less than was seen around the left temporal artery. The intima and media of the dissected vessel showed no inflammation or fibrinoid degeneration. Alcian blue and periodic acid-Schiff staining revealed no acid mucopolysaccharide.

Discussion

That the histologic picture of temporal artery disease is a classic one is emphasized in several of the reviews.[8,11] The accepted features of it, observed in the first biopsy in the case here presented, are (a) intimal proliferation and (b) inflammatory cellular infiltration of the adventitia and periadventitia (Figure 1). Two histologic features usually described in this disease, but not present in this biopsy specimen, are (a) degeneration of the inner elastica layer with giant cell formation and degeneration of the media of the vessel, and (b) narrowing of the lumen with thrombotic occlusion. Also atypical in the present case were laboratory studies which did not show the usual abnormalities observed in other cases.[8,11] The patient received prolonged glucocorticoid maintenance therapy (prednisone 30 mg daily from August 1965 through March 1966). Despite this large dose, symptoms referable to the opposite temporal region developed, leading to the excisional biopsy which showed a dissecting aneurysm. (Figure 3) The physical examination of the patient did not suggest any of the features associated with atypical Marfan's disease. Dissecting aneurysm of arteries other than the aorta was reviewed by Watson[9] and in none of the reports of 23 cases that he cited was a temporal artery involved. In the present case the dissecting episode could not be connected with history of trauma. The special histological stains (alcian blue and periodic acid-

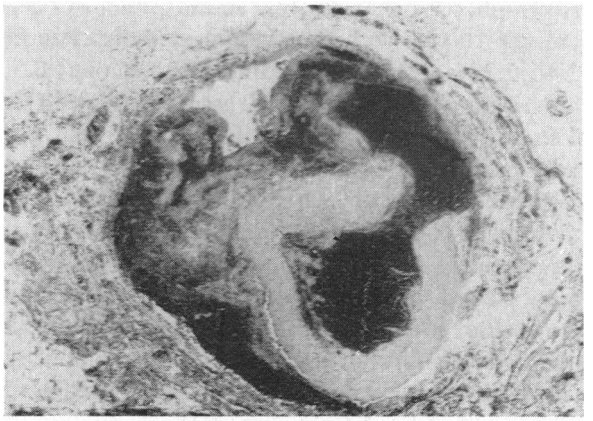

Figure 3.—Dissecting aneurysm of right temporal artery. Perianeurysmal adventitia shows some inflammatory reaction. (Hematoxylin and eosin stain, × 40.)

Schiff) showed no accumulation of mucoid-like material in the wall of the vessel such as is observed in idiopathic necrosis. Watson's classic definition of dissecting aneurysm is pertinent: "The lesion results from penetration of the blood into the wall of the artery, causing a concentric split along the natural lines of cleavage between coats or between medial laminae, extending over a variable distance and usually, but not necessarily, associated with a tear of the inner coat of the vessel."

The concomitant occurrence of dissection with long continued steroid therapy in the present case is interesting because of the reports of dissecting coronary artery aneurysms[5,10] in which the authors speculated upon the possibility of transitory lathyrism brought about by the changed hormonal status during pregnancy. It may be that in the case herein reported the large maintenance dose of prednisone suppressed the inflammation, since the cellular response to the arterial dissection was minimal (Figure 3) but the intra-arterial collagen tissue was altered ("loosened"[5]) sufficiently to facilitate dissection.

The cause of temporal arteritis is unknown. Its relationship to the autoimmune diseases is uncertain but there is a tendency to include temporal arteritis in the complex scheme of these disorders.[2] Giant cell aortitis[3] and temporal arteritis are considered related but knowledge of the basis of the relationship is sketchy.

The purpose of this communication is to add to the list of variables of temporal arteritis, hoping that widening the scope will, paradoxically, sharpen definitions.

Summary

Unusual manifestations in a case of bilateral temporal artery disease were onset at an earlier than usual age, incomplete histologic manifestations of temporal arteritis in the first biopsy site and recurrence in the opposite side with a histologic finding of dissecting aneurysm while the patient was receiving large amounts of glucocorticoid for maintenance therapy.

REFERENCES

1. Ainsworth, R. W., Gresham, G. A., and Balmforth, G. V.: Pathological changes in temporal arteries removed from unselected cadavers, J. Clin. Path., 14:115-119, 1961.

2. Alarcon-Segovia, D., and Brown, A. L.: Classification and etiologic aspects of necrotizing angiitides, Mayo Cl. Proceedings, 39:205-219, 1964.

3. Austen, G. W., and Blennerhassett, J. B.: Giant cell aortitis causing aneurysm of the ascending aorta and aortic regurgitation, New Eng. J. Med., 272:80-83, 1965.

4. Bethlenfalvay, N. C., and Nuslnowitz, M. L.: Temporal arteritis, Arch. of Int. Med., 114:487-489, 1964.

5. Brody, G. L., Burton, J. F., Zawadski, E. S., and French, J. A.: Dissecting aneurysm of the coronary artery, New Eng. J. Med., 273:1-6, 1965.

6. Das, A. K., and Laskin, D. M.: Temporal arteritis of the facial artery, J. of Oral Surg., 24:226-232, 1966.

7. Friedman, J. J.: Occult temporal arteritis, Am. J. Ophth., 60:333-335, 1965.

8. Noshay, W. C., and Roth, R. L.: Giant cell (temporal) arteritis, Henry Ford Hosp. Med. Bul., 10:455-461, 1962.

9. Watson, A. J.: Dissecting aneurysm of arteries other than the aorta, J. Path. Bact., 439-449, 1956.

10. Wells, A. L.: Dissecting aneurysm of coronary artery in peurperium, J. Path. Bact., 79:404-405, 1960.

Primary Malignant Mesothelioma of the Pericardium

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PRIMARY PERICARDIAL mesothelioma is rare. Only 29 cases had been reported to 1960,[7] with an additional eight cases through 1965. In virtually all of them diagnosis was made after death.*

So far as we could determine, the case described below is the first in which the diagnosis was established before death.

*Reference Nos. 2, 6, 11, 12, 15, 16, 20, 22.

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